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Unusual presentations of Creutzfeldt-Jakob disease: A case series and literature review

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Abstract

Transmissible spongiform encephalopathies are uncommon neurodegenerative diseases caused by misfolded prion protein deposition. Creutzfeldt-Jakob disease (CJD), the commonest human spongiform encephalopathy, has an average survival of 5 months. Common presenting symptoms are rapid cognitive decline, psychiatric disturbances, cerebellar ataxia, visual deficits, and movement disorders including myoclonus and parkinsonism. CJD may also present in atypical and nonspecific ways, hampering diagnosis. We present 3 pathologically verified cases with unusual onset and a review of recent literature on early clinical features of CJD. MRI and CSF biomarkers are generally less sensitive in patients with atypical presentations, and in such cases the disease course may be longer than in typical CJD.

Keywords: Unusual presentation, CJD, case series, literature review.

INTRODUCTION

Prion disease is a transmissible neurodegenerative disorder associated with deposition of misfolded prion protein in the brain. The commonest human form is Creutzfeldt-Jakob disease (CJD). About 85% of CJD is sporadic, with hereditary CJD making up a further 15%. Less than 1% of cases are classified as iatrogenic or variant CJD.[1,2] The annual incidence is 1-2 per-million[2], with a median survival of 5 months.[3] Other prion diseases include sporadic and familial fatal insomnia, Gerstmann-Straussler-Scheinker disease and Kuru disease.[1,4]

The prion protein consists of 210 amino acids, with three α helices and a short, antiparallel β pleated sheet. It occurs in 2 distinct conformers, type 1 and 2 (21 and 19 kDa respectively). The prion protein gene co-distributes with either methionine or valine at codon 129, which defines its genotype.[5] The protein is diglycosylated and anchored at the outer leaflet of the plasma membrane and serves multiple functions, including neuronal development, cell adhesion, axon guidance, synapse formation, ion homeostasis, myelin maintenance and neuroprotection.[4] In the disease state, the protein is misfolded into a β-sheet-rich conformation that is resistant to protease cleavage. It is amplified in neurons and astrocytes, and spreads from cell to cell through mechanisms that may involve exosomes, tunnelling nanotubes, retrograde and anterograde axonal transport and lysosomal exocytosis.[4] The abnormal protein aggregates in various parts of the body, but reaches high concentration in the central nervous system and the retina.[1,2] Intraneuronal accumulation of the misfolded protein leads to neuronal death, microglial activation, astrocytosis and spongiform vacuole formation, though the exact pathophysiology is poorly understood.[1,4] The incubation period varies from 1 to 42 years[2], depending on host prion protein genotype and exposure. The disease typically manifests as rapid cognitive decline, cerebellar ataxia, visual disturbance, pyramidal and extrapyramidal signs, and myoclonus, leading to death within months.[1] The clinical manifestations at symptom onset are highly variable, and this is reflected in the latest diagnostic criteria.[6] In contrast to previous criteria[7,8], these place less emphasis on onset symptoms and more on laboratory and MRI findings.[6,9] Initial clinical features, electroencephalogram (EEG) findings, MRI appearances and test characteristics of various biomarkers are highly variable and are modified

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by the prion protein codon 129 genotype[9], onset age[10], sex and molecular disease subtypes.[1] There are often delays in distinguishing CJD from a long list of disorders that can resemble it.[11,12] This will hamper patient selection for future therapeutic trials, several of which are now on the horizon.[11]

We describe a series of 3 patients with unusual onset clinical features of CJD, and survey recent literature on the atypical presentations of the condition. We discuss the application of the Braak hypothesis of topographic pathological progression to prion diseases, as advocated by Iwasaki.[13]

CASE REPORTS

Patient 1

A 63-year-old woman presented with a 6-month history of jerky vision that affected her ability to read and write. Examination showed ocular flutter. There was minimal difficulty with tandem walking without other cerebellar signs. Cognitive testing revealed a mild deficit; her Revised Addenbrooke's Cognitive Examination scored 93/100 (point deductions for memory and language, and greater weakness of verbal fluency). EEG, autoimmune screen, anti-neuronal antibodies and cerebrospinal fluid (CSF) 14-3-3 protein done within 5 months of disease onset were all normal/negative, with the exception of a weakly positive anti-CV2 antibody. Brain magnetic resonance imaging (MRI-B) showed subtle cortical ribboning in the left parietal and temporal regions. A diagnosis was established by brain biopsy, which demonstrated spongiform encephalopathy and prion protein immunoreactivity typical of CJD.

Her disability progressed with worsening ocular flutter, ataxia, myoclonus, dysarthria and cognitive decline with psychosis. She died 2 years after symptom onset.

Patient 2

This 73-year-old man complained of constant high-pitched tinnitus, which began subacutely following a viral prodrome. Audiometry showed mild low frequency sensorineural hearing loss on the left. There were no other cranial nerve, pyramidal or cerebellar signs. His stance and gait were normal.

Investigations, including serum anti-neuronal antibodies, CT brain, MRI brain and CSF analysis were unremarkable, though CSF 14-3-3 protein testing was not done.

Two months later, he presented with 2 episodes of syncope and complained of gait unsteadiness and intermittent paraesthesias on his torso. Gait ataxia progressed rapidly over the next month, postural stability was impaired, and he had a number of falls. Three months after presentation, his family noticed progressive cognitive impairment. A trial of levodopa therapy produced no benefit.

Six months after the initial presentation he was admitted to hospital with severe gait ataxia. He required a frame to mobilise and his Montreal Cognitive Examination score was 24+1/30. Repeat MRI brain and spine was unrevealing, and an EEG showed only mild generalized theta-delta slow waves. He died 13 months after symptom onset. The diagnosis of CJD was established on autopsy.

Patient 3

At the age of 38, this man presented with 6 months of sleep disturbance. He could only obtain 2-3 hours of disrupted sleep at night. His wife observed thrashing activity and excessive snoring in sleep, with frequent sleep arousals. He would often be confused and disorientated for some minutes after waking. There were subtle memory and attentional deficits, and he had been unable to continue his employment.

Examination showed subtle dysarthria. Incoordination with dysdiadochokinesis was present in his right arm and both legs. He walked unsteadily. His gait pattern was unusual, with staggering and high steppage features.

MR brain and spine with contrast were normal. A standard EEG recording was dominated by multiple cycles of sleep onset and arousal, which was accompanied by kicking lower limb movements. A sleep study showed severe sleep fragmentation with frequent arousals and prolonged wakefulness. CSF 14-3-3 and real-time quaking induced conversion test (RT-QUIC) were negative 9 months after presentation. Sequencing of the PNRP (prion protein) gene did not detect pathogenic mutations. FDG-PET scan showed bilateral thalamic hypometabolism. A diagnosis of sporadic fatal insomnia was made. His sleep pattern improved marginally with olanzapine and sodium valproate. Cognitive function steadily declined. He was non-communicative at 18 months and died 25 months after presentation. Autopsy revealed findings of spongiform encephalopathy in keeping with CJD. In addition, the fatal insomnia pattern of severe bilateral thalamic neuronal loss was present.

DISCUSSION

CJD typically presents with prodromal changes in mood and behavioural plus subtle cognitive decline.[6,11] In a survey of 492 patients with probable and definite CJD, the commonest initial clinical features were dementia (37%), cerebellar ataxia (34%), visual symptoms (15%) and psychiatric disturbance (14%). About 10% had nonspecific complaints such as headache, fatigue, sleep disturbances and "peculiar feeling in the head".[14]

The atypical early features of CJD are well summarised by Kasikaki *et al*.[15] and Baiardi S *et al*.[16] They describe neuropsychiatric symptoms—mild behavioural disturbances, mood disorder, conversion disorder, personality changes, psychosis, delusions[17] and hallucinations.[18] Patients presenting with visual symptoms (including blurring, reduced acuity, impaired colour vision, metamorphopsia, visual snow, visual neglect, abnormal visuospatial perception, visual agnosia, diplopia, visual field defect and cortical blindness) are classified as having the Heidenhain variant, which accounts for 3.7-10% of CJD.

Although more common in the advanced stages, focal seizures and myoclonus including status epilepticus[19] may infrequently occur at presentation.[16] Other uncommon presenting features include movement disorders such as parkinsonism[20,21], corticobasal syndrome, progressive supranuclear palsy-like or multisystem atrophy-like states[22], focal or generalised dystonia, chorea, athetosis, blepharospasm, isolated tremor, alien hand syndrome, mirror movements[23] and hemiballismus.[15,16] There are rare case reports of isolated aphasia[24], apraxia[25], dysexecutive syndrome[26], acoustic agnosia, hearing loss, acute stroke-like syndrome of monoparesis or hemiparesis, isolated brainstem syndrome[27], peripheral neuropathy[15] and amyotrophy.[28]

MRI is a key investigation. In typical CJD, the findings are restricted diffusion in at least 2 cortical regions, which shows up as "cortical ribbon" sign; or restricted diffusion in the caudate nucleus, putamen and thalamus. The overall sensitivity varies between 80-98% and specificity between 92-97%, depending on the scanner, study focus and protocols.[9] CSF 14-3-3 protein has an overall sensitivity of 92%, but lower in early stage of the disease and in MV2 and MM2 genotypes (60-70%); and specificity of 40-92%.[9] CSF RT-QuIC has a sensitivity of 92-97% but lower in MV2 (75-93%), VV1 and MM2 (44-78%) subtypes, especially MM2 thalamic subtype which presents a sporadic fatal insomnia.[9] A meta-analysis of CSF markers showed the overall sensitivity of RT-QuIC to be 89.5% with near 100% specificity, and those of 14-3-3 protein to be 87.1% and 90.2% respectively.[29] As detailed in a recent review, there is yet no systematic study of the sensitivity and specificity of FDG-PET in CJD.[30,31]

All three of our patients had definite CJD on histopathology. All had unusual onset symptoms and relatively prolonged survival. Patient 1 developed ocular flutter, which is associated with lesion in either the paramedian pontine reticular formation[32] or the dorsal vermis/fastigial nucleus of the cerebellum.[33] Patient 2 presented with bilateral high frequency tinnitus for 2 months before onset of gait ataxia and cognitive decline. High frequency constant bilateral tinnitus is hard to localise, but may have been caused by damage to the primary auditory cortex.[34] Patient 3 presented with severe sleep fragmentation and FDG-PET scan suggested early bilateral thalamic involvement. MRI brain at presentation showed typical CJD change only in Patient 1, and even then only in one region, and thus did not fulfill the MRI diagnostic criteria.[9] CSF 14-3-3 protein was tested in Patients 1 and 3, and the RT-QuIC in Patient 3; all were negative despite symptomatic disease. This led to delayed diagnosis in these patients.

In a multinational survey of 2,304 sporadic CJD patients in Europe and Australia, the median survival was reported to be 5 months.[3] A survey in China of 104 patients reported median survival of 9 months[35], and a third survey of 400 patients in Taiwan documented a median survival of 10 months.[36]

Our Patients 1 and 3 survived for more than 2 years and Patient 2 for 13 months, longer than the median survival reported in the literature.[11,35,37,38] Longer survival is reported to be associated with younger onset age, female gender, codon 129 heterozygosity, presence of CSF 14-3-3 protein, type 2a prion protein[3], absence of seizures and periodic sharp wave complexes[36], and lower CSF tau protein level.[37] Prolonged survival of Patients 1 and 3 could have been due to lead time effect as the anatomical sites of initial disease involvement were symptomatic.

Braak *et al*'s widely accepted staging system for Parkinson's disease proposes a non-random progressive caudal to rostral spreading of alpha-synuclein immunostaining pathology, beginning in the dorsal motor nucleus of the vagus nerve in the medulla and in the anterior

olfactory nucleus, before ascending to the cerebral neocortex.[39] A growing body of evidence suggests that alpha-synuclein protein is able to spread transcellularly and induce pathological aggregation in a prion-like manner.[40] CJD is the archetypal prion disease, and Iwasaki proposed that CJD also progresses topographically, though with a different pathological onset region in each individual and from thence, spreading without the directionality or hierarchy of Braak's scheme for Lewy pathology.[13] In our first patient, the progression from ocular flutter to ataxia and then cognitive deficits suggests that the disease progressed from brainstem or vermis/fastigial nucleus of the cerebellum to the greater cerebellum and to the cerebral cortex. Patient 2 had probable temporal cortical disease at onset, spreading to the cerebellum or cerebellar connection centres and the wider cerebral cortex. In Patient 3, it is probable that the disease progressed from the bilateral thalami to the cerebral cortex.

The utility of Iwasaki's proposal is in alerting the clinician of the possible diagnosis of CJD in patients with atypical onset symptoms, which can be highly variable, and as illustrated in our cases, often have negative investigation findings. Examples of a wide range of atypical onset symptoms in CJD abound in the literature[15,18–20,22–28,41–47], and the list of differential diagnoses is long.[12] The subacute onset but relentlessly progressive involvement of different neurological systems is the common thread that ties the bewildering array of onset clinical features to the diagnosis of CJD.

DISCLOSURE

Ethics: Written consent from the next-of-kins of all subjects has been obtained.

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